

Coeliac disease in children of West Indian origin

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Abstract

Coeliac disease is uncommon in populations of non-European origin. Two English born West Indian children with coeliac disease are presented. The diagnosis should be considered in children of West Indian origin with chronic diarrhoea.

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Keywords: coeliac disease, West Indian children, HLA phenotypes.

The development of coeliac disease requires exposure to gluten and it is strongly influenced by genetic factors. Coeliac disease, that is, permanent gluten intolerance, predominantly affects populations of European origin. There are now reports of the disease in children from the Indian subcontinent, Lebanon, Iraq, Kuwait, Sudan, and Cuba but it appears to be very rare.¹⁻⁶ Presentation in these patients is generally delayed to the second or third year of life. This may be due to prolonged breast feeding and late introduction of gluten in their diet.

Transient gluten intolerance has been described in children presenting at less than 2 years of age who eventually return to a normal diet.⁷ It is important to distinguish it from coeliac disease to avoid overdiagnosis, especially in the low risk population.

We report coeliac disease in two English born children of West Indian origin.

Case reports

CASE KM

This girl was initially seen at 22 months of age for poor feeding, chronic diarrhoea for two months, protuberant abdomen, and irritability. Her blood film showed microcytosis and hypochromia. Low serum iron and ferritin (7 mmol/l and <5 mg/l) confirmed the presence of iron deficiency. Her small intestinal histology showed moderate to severe villous atrophy with increased intraepithelial lymphocytes and crypt hyperplasia. Gluten-free diet was advised. Due to social circumstances, she did not keep to her exclusion diet and failed to attend follow up appointments for 15 months.

She was seen again aged 3.8 years when her gliadin, reticulin, and endomysial antibodies were strongly positive on a gluten containing diet. She was still iron deficient (ferritin <5 mg/l) and her stool culture was negative for pathogens. Small bowel biopsy was repeated and the histology showed an almost flat mucosa with crypt hyperplasia and increased intraepithelial lymphocytes. Advice was given again that she should return to the gluten-free diet. Subsequent review found that she had

maintained her gluten exclusion diet and showed good weight increment. Her serum iron and ferritin had improved (9.2 mmol/l and 20 mg/l) and her gliadin, reticulin, and endomysial antibodies were all negative.

Both her parents were black Jamaicans. However, her maternal great great grandmother was white. Class II HLA typing showed that she was positive for DQw2, DR3, and DRw52.

Although this case does not fulfil the original guidelines of the European Society of Paediatric Gastroenterology and Nutrition (ESPGAN),⁸ it satisfies the later revised criteria for the diagnosis of coeliac disease.⁹

CASE WP

This boy presented in 1974, aged 6 months, with failure to thrive and persistent diarrhoea. Small bowel histology showed a flat mucosa with severe villous atrophy and crypt hyperplasia. He was started on a gluten-free diet and he began to thrive. A pre-gluten challenge small bowel biopsy was normal at three years of age. He was then challenged with gluten for three months. His post-gluten challenge biopsy showed only mild villous atrophy. He remained on a gluten containing diet and continued to thrive.

Another small bowel biopsy was carried out at 4.5 years of age and it appeared normal. At that stage, he was thought to have transient gluten intolerance.

He was monitored from the age of 6 years for gliadin and reticulin antibodies. Both antibodies became positive at 9 years of age and, although he remained well and thriving, a small intestinal biopsy was repeated. On this occasion, it showed a flat mucosa with severe villous atrophy and crypt hyperplasia. This confirmed the diagnosis of coeliac disease and he was advised to return to gluten-free diet. He remained well on subsequent review.

His mother was white and his father was a black Jamaican. HLA class II typing showed that he was positive for DQw3, DR4, DRw52, and DRw53.

Discussion

Although histology is crucial for the diagnosis, gliadin, endomysium, and reticulin antibodies have been shown to be very useful adjuncts for diagnosing and monitoring coeliac disease.¹⁰ As in patient WP, who was mentioned in a previous paper,⁷ histological relapse was heralded by the appearance of these antibodies and during that time he was in good health. This case illustrates the importance of long term follow up in children with the presumed

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diagnosis of transient gluten intolerance. Indeed, it is difficult to make a final diagnosis of transient gluten intolerance, as children may take many years to relapse.⁷

Both children are a mixture of black West Indian and white ethnic backgrounds. The first case, with a distant Scottish relative, showed a typical presentation and response to gluten provocation, although the diagnosis was complicated by social circumstances. In contrast, patient WP, with mixed parentage, had a greatly delayed response to dietary challenge, only relapsing after six years of exposure to gluten. The HLA status may be relevant, being the commonly associated pattern in patient KM, but of less common association in patient WP. The association of coeliac disease with DR4 antigen has been reported in only a small number of patients, and different gene alleles are believed to be responsible.^{11 12} Furthermore, the degree of gluten sensitivity may be different in DR3/DQw2 negative patients as their anti gliadin antibody titres are lower than in the DR3/DQw2 positive patients.¹³

The lack of reports of coeliac disease in children of West Indian origin may be partly due to low gluten consumption in the West Indies. However, the situation may change when they move to another country such as the United Kingdom where gluten consumption is considerably higher. The HLA-DR3 and HLA-DQw2 antigens^{12 14 15} have a stronger association with coeliac disease than HLA-B8 antigen which has a low prevalence in the West Indian population.¹⁶ As both of our patients possess class II HLA markers (DR3-DQw2 and DR4¹⁴) for coeliac disease, it would be interesting to find out the prevalence of these antigens in the West Indies. It is also possible that the presence of coeliac disease in these cases is due to their mixed ancestry and it may not develop in West Indians of black African origin.

In conclusion, despite the uncommon occurrence of coeliac disease in the West Indies, the diagnosis should still be considered in children of West Indian origins with chronic diarrhoea.

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